A reliability study of impairment and disability scales for myasthenia gravis patients

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Summary

The authors developed two scales to be adopted for the evaluation of myasthenia gravis (MG) patients. The first scale (MG impairment scale) is based on objective patient evaluation and on patients' responses to standardized questions relating to the functioning of specific muscle groups. It consists of 13 items exploring strength and 10 items exploring fatigability. The second scale (MG disability scale) evaluates disability in those everyday activities that are often impaired in MG patients. Test-retest reliability of each item and of the global score (sum of single item scores) was assessed by the weighted K statistic and by the intraclass correlation coefficient. Reliability was invariably 'substantial', and for single items 'almost perfect' for the MG impairment scale, and invariably 'almost perfect' for the MG disability scale. The internal structure of the MG impairment scale was explored by means of the principal component analysis. This analysis resulted in three main (rotated) factors, which loaded respectively onto 'ocular', 'spinal' and 'bulbar' functions. For these factors, we report factor score coefficients that can be used to compute single patients' scores, which in turn may be used in further analyses, particularly for follow-up studies. We also report the results of an analysis of the correlations between the two scales. The MG impairment and the MG disability scales are proposed for application in both clinical and research settings.

KEY WORDS: Assessment, disability, impairment, myasthenia gravis, reliability, scales.

Introduction

Myasthenia gravis (MG) is caused by an immune-mediated involvement of the neuromuscular junction, and is characterized by muscle weakness and fatigability. Since the symptoms and signs of MG fluctuate, it can be very difficult to evaluate with precision a patient's clinical condition at any given point. A careful patient assessment should consider both weakness and fatigability in key muscular districts; this is usually relatively easy in spinal muscles, but often difficult in bulbar districts. A reliable assessment of the latter additionally requires both direct and indirect recording of global functions, like speech and swallowing (and the degree of their involvement). A scale for the clinical evaluation of MG patients should be sensitive enough to allow, in the daily management of patients, recognition and measurement of even slight changes, such as those caused by the 'end of dose' effect of the anticholinesterase drug. Assessment should also be quick to perform. In view of all these requirements, the need for and usefulness of a reliable scale are probably more crucial in MG than in any other neurological disease. Patient management apart, a reliable scale is particularly valuable for conducting clinical trials. But, as is the case with other clinical contexts, no single instrument is likely to suit all clinical and research settings, as the focus of specific interest may vary considerably, ranging, for instance, from assessment of quality-of-life changes after a given medical intervention to bedside evaluation of minor neurological changes; moreover, simple linear relationships between such different levels of assessment are unlikely – for instance, a worsening in extraocular muscle impairment may paradoxically relieve diplopia-related disability.

In a recent paper (1) published by an *ad hoc* Myasthenia Gravis Foundation of America task force, it was suggested that the evaluation of an MG patient should be a multidimensional assessment based on a clinical classification, a quantitative MG score, a therapy status, a post-intervention status and a morbidity and mortality status. The authors underline that their proposed instruments will probably undergo amendments and refinements, and they recommend the development of other instruments for the exploration of different dimensions, particularly quality of life. To date, the most widely adopted MG scale is a modification of Osserman's (2) clinical classification (3), which consists simply of five levels that define the extent of the neurological involvement (ocular, spinal, bulbar), and distinguishes categories on the basis of disease course. However, within the single levels established by this classification, the degree of impairment and of disability may vary considerably. This scale, as well as the more uniformly structured clinical classification proposed by Jaretzki et al. (1), may be useful for the identification of subgroups of patients, but not for their longitudinal monitoring. A widely used scoring system, which can also be adopted retrospectively, and whose six levels reflect degree of disability regardless of the systems actually involved, was

introduced by Oosterhuis et al. (4), but never adequately validated. Szobor's (5) scale, overtly derived from Kurzke's Disability Status Scale (6), includes some items (like 'psychic fatigue') that are not really appropriate to myasthenia, and the scale's reliability has been never ascertained.
Other clinical scales have been proposed, but most of them have been used only sporadically and are poorly validated. The scale proposed by Besinger et al. (7) is based both on objective clinical evaluation of single districts and on some patient-reported information. The diffusion of this scale was probably hampered by the need for certain instrumental devices (dynamometer, spirometer). The MG scale of Tindall et al. (8), derived from the previous one, also requires some equipment. The reliability of a modified version of the Tindall et al. scale (8) was recently tested (9) on a small number of subjects (five patients and four normal subjects) for whom the pooled standard deviation of the global score was estimated; this was the scale used to calculate the quantitative MG score included in the Myasthenia Gravis Foundation of America paper (1). A further study assessing the reliability of the original Tindall et al. scale (8) and that of a simple 9-item scale developed and used by Gajdos et al. (10) has been recently published (11). The authors conclude that both scales are highly reliable and that correlation between them is high. D'Alessandro et al. (12) proposed an eight-item scale based on standardized questions and on patient observation. They tested its inter- and intra-rater reliability on a small (no.=12) patient sample, and found that for each item the reliability score was higher than that of Osserman's scale (3). The relationship between Tindall's modified quantitative MG score (9) and a newly developed questionnaire assessing items that investigate the impact of MG symptoms on activities of daily living was recently investigated by Wolfe et al. (13), who found a significant correlation coefficient between the two global scores. Although of some practical interest, this observation may constitute an oversimplification of the complex relationships between the objective neurological picture and its consequences on the patient's wellbeing and social functioning. Finally, some scales have been proposed that may find application in limited settings. Schumm and Dichgans (14) developed a rating scale for ocular symptoms; Nathadwarawala et al. (15) a test for swallowing capacity and Nicklin et al. (16) for shoulder abduction fatigability. These instruments may be valuable in limited clinical and research settings, but their usefulness in routine MG patient global assessment is questionable.
In the present study, we developed a new scale, measuring strength and function in key districts, as well as fatigability, to be used as a routine instrument in the bedside assessment of MG patients (MG impairment scale). For this purpose, the scale was designed to be more analytical and comprehensive than existing ones. Through their application to a considerable number of patients, its items have been found to show both high sensitivity, i.e., a high capacity to detect subtle changes in MG impairment, and high inter-rater reliability. At the same time we developed a second score, which assesses the patient's disability in activities of daily living (MG disability scale). Its test-retest reliability was also assessed, as was the relationship between this scale and the MG impairment scale.

Materials and methods

MG impairment scale

The MG impairment scale (Appendix) consists of 13 items measuring the strength of specific muscle groups and 10 items evaluating fatigability. Strength is assessed in accordance with the MRC criteria, and additionally, in selected items, evaluates more global functions (specifically the ability to move from the supine to the sitting position, ptosis and diplopia). With the exception of the item 'diplopia', this part of the scale is based completely on objective patient testing. The assessment of fatigability, on the other hand, is based both on patient examination and on standardized questions relating to common activities involving specific muscle groups. The order of presentation of the single 'strength/function' and 'fatigability/function' items was standardized and is reported in Table 1. The present version of the MG impairment scale is the result of a series of refinements and of preliminary test-retest reliability assessments, which involved testing 60 MG patients and 20 normal subjects.

MG disability scale

The MG disability scale (Appendix) consists of 6 items exploring the main causes of disability in everyday activities. For each item, the patient has to select the level (out of a total of four) that most accurately reflects his situation in the previous 24 hours.

Reliability assessment

Thirty patients (17 males) were selected for the reliability study. They were affected by MG, diagnosed according to clinical and electrophysiological criteria. Their age ranged from 18 to 78 years; 83% had a positive antireceptor antibody test.
The reliability study was performed by three examiners, two neurologists and one resident physician at the School of Neurology. The three examiners formed 3 pairs (AB, AC, BC) and each pair tested 10 MG patients: thus, each examiner performed a total of 20 tests. Each single test involved two assessments separated by a 10-minute interval. The order in which each member of the pair of examiners tested the patient was randomized. Moreover, in each group of 10 patients, we included at least 6 with ocular symptoms, at least 6 with spinal and at least 6 with bulbar symptoms. The two neurological assessments were performed in the morning and were respectively scheduled for 90 and 130 min after anticholinesterase ingestion, on the assumption that the clinical effect of the drug between the two evaluations was steady.
Before each of the two assessments, each patient had to answer the questions of the MG disability scale questionnaire presented by the two examiners.

Statistical analysis

Systematic test-retest effects (i.e., the possible influence of fatigue on the second examination) and effects

Table I - MG impairment scale: absolute frequency of scores in the 23 items (60 assessments).

Order*	Item	Level 0	Level 1	Level 2	Level 3
	Strength / function				
1	Sitting from supine	22	15	10	13
2	Psoas	10	12	15	23
7	Ptosis	22	13	14	11
9	Oculomotor paresis	22	14	12	12
10	Diplopia	20	12	14	14
11	Orbicularis oculi	18	22	12	8
12	Facialis inferior	21	11	13	15
13	Tongue	23	13	12	12
14	Masseter	27	12	13	8
21	Neck flexors	28	11	13	8
6	Finger radial extensors	18	15	14	13
5	Triceps	19	17	15	9
4	Deltoid	24	12	12	12
	Fatigability / function				
8	Ptosis	25	18	17	
15	Upper limb girdle	27	13	8	12
3	Lower limb girdle	18	14	15	13
16	Chewing	30	12	11	7
17	Tongue	18	42		
18	Swallowing	25	12	18	5
19	Voice	14	13	14	19
20	Breathing	35	12	13	0
22	Neck flexors	23	24	13	
23	Squats	16	12	14	18

* Order of execution of the single items.

related to each single examiner were assessed by logistic regression analysis for each item of both the MG impairment and the MG disability scale, as well as on their global scores. The global score was the sum of the single item scores.

Interobserver agreement (reliability) was assessed by computation of the weighted K statistic in accordance with Fleiss (17,18), the results being interpreted on the basis of the usual categories: 0-0.2 = slight; 0.21-0.4 = fair; 0.41-0.6 moderate; 0.61-0.8 = substantial; > 0.8 = almost perfect agreement. The reliability of the global scores was assessed by the intraclass correlation coefficient (ICC) (19).

The internal structure of the MG impairment scale was explored by means of a principal component analysis (PCA) on the item scores; both the unrotated solution and the varimax rotated solutions were taken into account. The clinical meaning of the PCA factors was assessed by analysis of the association between impairment scale items and PCA factors. For this purpose non-parametric Spearman correlation coefficients were computed. When a statistically satisfactory and clinically meaningful solution of PCA analysis is found, main PCA factors can be considered as synthetic indices of underlying independent dimensions, each of which is originally measured by a series of correlated items. Each single patient's factor scores can be computed as follows and used in further analyses:

FSxn = V1n x FSc1 + V2n x FSc2 +...+ Vzn x FScz, in which FSxn is the factor score for the x th factor of the n th patient; Vznx is the value of the z th variable of the n th patient and FScz is the factor score coefficient for the z th factor.

Results

Logistic regression analysis did not demonstrate any significant effect related to the order of testing (first/second examination), nor to the specific examiner.

The absolute frequencies of the scores obtained in the 23 items of the MG impairment scale are reported in Table I. All the levels of all the items, with the exception of breathing, appeared to be well represented in our patient sample.

Table II (for tables II-V, see over) reports the K statistic for the 23 items of the MG impairment scale. All the items showed at least 'substantial' reliability. The ICC of the global MG impairment scale was R = 0.93, which means 'almost perfect' reliability.

The absolute frequencies of the MG disability scale scores are reported in Table III, and the inter-rater reliability statistics for the single items in Table IV. For each item, reliability was 'almost perfect'. Likewise, the global MG disability scale ICC showed 'almost perfect' reliability (R = 0.95).

The results of the PCA performed on the items of the MG impairment scale are shown in Table V. The analysis extracted 3 factors with eigenvalues greater than 1, which explained 73% of the global variance. The first factor in the unrotated solution correlates with almost all the origi-

nal items, and can be considered and used as a general index of disease severity. Regarding the varimax rotated solution, the clinical meaning of the three factors can be seen easily in Table V, which reports the Spearman correlation coefficients between the original items and the factor scores: factor 1 is clearly an 'ocular' factor, with some minor relation to facial muscle strength; the second factor shows high loading on spinal muscular function, and the third correlates mostly with bulbar functions.
The correlation coefficients between MG disability scale items and MG impairment factor scores are reported in Table VI. The first unrotated impairment scale factor correlates with almost all items of the disability scale, with the exception of 'breathing' (no correlation) and 'voice' (minor significance). The first rotated, 'ocular' impairment factor correlates almost exclusively with the disability scale item 'vision'; the 'spinal' impairment factor correlates strongly with limb disability, but also with some 'bulbar' disability items; the bulbar impairment factor is strongly associated with bulbar disability items and with respiration. Table VI (see p. 142) also reports the factor score coefficients, which can be used as explained in the methods section.

Table II - MG impairment scale: inter-rater reliability of the 23 items.

Item	Observed agreement	Expected agreement	K	z
Strength / function				
Sitting from supine	0.90	0.27	0.86	7.9
Psoas	0.70	0.28	0.61	5.4
Ptosis	0.80	0.26	0.73	6.9
Oculomotor paresis	0.70	0.25	0.64	6.5
Diplopia	0.69	0.24	0.61	5.5
Orbicularis oculi	0.77	0.28	0.67	6.1
Facialis inferior	0.77	0.26	0.68	6.4
Tongue	0.80	0.27	0.73	6.9
Masseter	0.73	0.31	0.61	5.5
Neck flexors	0.77	0.32	0.66	5.9
Finger radial extensors	0.80	0.25	0.73	6.9
Triceps	0.80	0.26	0.73	6.9
Deltoid	0.63	0.28	0.63	5.8
Fatigability / function				
Ptosis	0.87	0.34	0.80	6.1
Upper limb girdle	0.84	0.56	0.67	7.2
Lower limb girdle	0.88	0.51	0.63	5.0
Chewing	0.76	0.33	0.62	5.0
Tongue	0.84	0.58	0.61	2.7
Swallowing	0.78	0.31	0.63	5.0
Voice	0.73	0.20	0.67	7.3
Breathing	0.80	0.43	0.65	4.9
Neck flexors	0.80	0.35	0.69	5.3
Squats	0.73	0.21	0.66	7.2

Table III - MG disability scale; absolute frequency of scores in the 6 items (60 assessments).

Item	Level 0	Level 1	Level 2	Level 3
Upper limbs	26	17	13	4
Lower limbs	24	12	16	8
Eating	22	15	17	6
Voice	14	15	25	6
Sight	18	16	16	10
Breathing	36	16	8	0

Discussion

Compared to existing scales, our MG impairment scale deepens analysis through the inclusion of a greater number of items. This analytical depth may be particularly valuable for its application in clinical settings. The time needed for application (about 15-20 minutes) is entirely reasonable. From the point of view of research purposes, each single item seems satisfactorily reliable, and the global score shows 'almost perfect' reliability. Unlike other 'objective' scales, ours avoids the use of technical equipment for the assessment of strength and of specific functions, as we believe that even if this approach is able to improve sensitivity, longitudinal reliability (patients are often followed up for very long periods) and between-center transferability may not be optimal. The internal

structure of the MG impairment scale was analyzed by PCA. This analysis explores the global set of data computing new 'factors' that are weighted sums of the original variables. These factors are, by definition and in contrast to the original variables, independent (uncorrelated) and are arranged (in decreasing order) according to the amount of the global variance they account for ('explain'). Typically the first few factors (depending on the number of the original variables) explain a high percentage of the variance, meaning that the other ones can be safely ignored without losing important information about the global structure. The analysis of the association (correlation) between each new factor and the entire set of the original variables is the key for its interpretation. A PCA is to be considered valuable if this interpretation of the factors proves to be clinically meaningful. PCA is thus a statistical method that analyzes the structure of the data, as well as a method of data reduction because the number of important factors (in terms of explained variance) is far lower than the number of the original variables. The results of the application of this analysis to our data set was consistent, in that the meaning of the main factors had clinical importance. Moreover, it demonstrated that strength and fatigability are strongly

Table IV - MG disability scale: Inter-rater reliability.

Item	Observed agreement	Expected agreement	K	z
Upper limbs	0.97	0.32	0.95	8.1
Lower limbs	1	0.29	1	9.0
Eating	0.97	0.29	0.95	8.5
Voice	0.90	0.30	0.86	7.6
Sight	0.93	0.26	0.91	8.5
Breathing	0.93	0.45	0.88	6.3

Table V - MG impairment scale: Spearman correlation coefficients between PCA factors and original item scores / factor score coefficients.

Item	NR 1	R 1	R 2	R 3
Strength / function				
Sitting from supine	0.62 / 0.041	0.22 / 0.005	0.12 / 0.002	0.33 / 0.010
Psoas	0.75 / 0.052	0.05 / 0.000	0.76 / 0.054	0.32 / 0.010
Ptosis	0.72 / 0.050	0.81 / 0.056	0.26 / 0.006	0.21 / 0.003
Oculomotor paresis	0.71 / 0.049	0.74 / 0.051	0.23 / 0.005	0.21 / 0.003
Diplopia	0.57 / 0.040	0.69 / 0.048	0.21 / 0.004	0.36 / 0.012
Orbicolaris oculi	0.67 / 0.048	0.67 / 0.046	0.56 / 0.042	0.51 / 0.040
Facialis inferior	0.64 / 0.043	0.55 / 0.041	0.67 / 0.045	0.44 / 0.020
Tongue	0.85 / 0.067	0.03 / 0.000	0.23 / 0.005	0.82 / 0.068
Masseter	0.65 / 0.044	0.12 / 0.001	0.32 / 0.010	0.65 / 0.046
Neck flexors	0.77 / 0.053	0.31 / 0.009	0.67 / 0.045	0.55 / 0.042
Finger radial extensors	0.84 / 0.063	0.21 /0.004	0.88 / 0.072	0.25 / 0.007
Triceps	0.81 / 0.056	0.09 / 0.007	0.79 / 0.056	0.41 / 0.015
Deltoid	0.91 / 0.071	0.33 / 0.010	0.73 / 0.051	0.55 / 0.043
Fatigability / function				
Ptosis	0.74 / 0.052	0.67 / 0.046	0.34 / 0.012	0.31 / 0.010
Upper limb girdle	0.72 / 0.049	0.12 / 0.001	0.78 / 0.055	0.20 / 0.003
Lower limb girdle	0.69 / 0.048	0.23 / 0.005	0.75 / 0.053	0.22 / 0.004
Chewing	0.81 / 0.056	0.03 / 0.000	0.31 / 0.009	0.75 / 0.054
Tongue	0.75 / 0.051	0.11 / 0.001	0.12 / 0.002	0.88 / 0.071
Swallowing	0.67 / 0.048	0.24 / 0.005	0.21 / 0.003	0.79 / 0.056
Voice	0.73 / 0.049	0.33 / 0.010	0.55 / 0.041	0.68 / 0.047
Breathing	0.53 / 0.038	0.23 / 0.005	0.21 / 0.003	0.65 / 0.046
Neck flexors	0.45 / 0.032	0.22 / 0.004	0.60 / 0.042	0.54 / 0.042
Squats	0.66 / 0.048	0.32 / 0.009	0.78 / 0.055	0.34 /0.015

Abbreviations: NR: non rotated solution; R: varimax rotated solution.

Table VI - Spearman correlation coefficients between MG disability scores and MG impairment scale factor scores.

Item	NR 1	R 1	R 2	R 3
Upper limbs	0.54	0.12	0.62	0.45
Lower limbs	0.52	0.32	0.58	0.41
Eating	0.67	0.25	0.55	0.80
Voice	0.33	0.34	0.54	0.72
Sight	0.60	0.55	0.33	0.26
Breathing	0.23	0.24	0.32	0.67

Abbreviations: NR: non rotated solution; R: varimax rotated solution.

related within single anatomo-functional districts (ocular/spinal/bulbar), but not between differing ones. The factor score coefficients here reported can be used to compute synthetic indices of 'ocular', 'spinal' and 'bulbar' scores for single patients. These scores are particularly useful in follow-up studies and in clinical trials.
The MG disability scale is more similar to existing scales, which explore patient-reported functions (e.g. 12). However, with our disability scale, single item reliability is even higher, probably because the score is assigned by the patient himself in response to the examiner's questions. The single items of the disability scale show congruent correlations with the PCA factors, a fact that increases confidence in its consistency. The disability scale can be proposed as an instrument to monitor large numbers of patients, when objective evaluation is not feasible.
We believe that our scales constitute easily applicable and reliable instruments that may improve the assessment of impairment and disability in myasthenic patients.

Appendix

MG impairment scale

A) STRENGTH / FUNCTION

I ***Sitting from supine***
0 Sits easily from supine
1 Sits from supine with arms outstretched
2 Sits from supine with examiner holding his legs
3 Does not sit from supine without aid

II ***Psoas*** *(evaluation in sitting position)*
0 Strength 5
1 Strength >= 4
2 Strength >= 3
3 Strength <3

III ***Ptosis***
0 No ptosis
1 Ptosis at upper margin of pupil
2 Ptosis covering half pupil
3 Complete ptosis

IV ***Oculomotor paresis*** *(6 eye positions must be checked and only evident paresis should be counted)*
0 Absence of oculomotor paresis
1 Paresis of 1 - 2 muscles
2 Paresis of 3 - 4 muscles
3 Paresis of more than 4 muscles

V ***Diplopia*** *(check 6 gaze positions plus primary position)*
0 No diplopia
1 Diplopia in 1 gaze position
2 Diplopia in 2 gaze positions
3 Diplopia in 3 or more gaze positions

VI ***Orbicularis oculi***
0 Strength 5 (closes eyelids with normal force)
1 Strength > = 4 (closes eyelids with slightly reduced for - ce)
2 Strength > = 3 (closes eyelids completely, but with mini - mal force)
3 Strength <3 (lagophthalmus)

VII ***Facialis inferior*** *(examine the ability to protrude lips, smile forcefully and blow against resistance)*
0 Strength 5 (performs the tasks with normal strength)
1 Strength >=4 (slight decrease of strength in performing the tasks)
2 Strength >=3 (moderate decrease of strength in perfor - ming the tasks)
3 Strength <3 (performs the tasks with minimal strength or unable to perform at least one of the tasks)

VIII ***Tongue***
0 Normal protrusion and normal strength against the cheeks
1 Slight reduction of protrusion and/or of strength against the cheeks
2 Poor protrusion and does not move the cheek
3 No or minimal movements

IX ***Masseter***
0 Strength 5
1 Strength >=4
2 Strength >=3
3 Strength <3

X ***Neck flexors*** *(patient supine, with abducted arms)*
0 Strength 5
1 Strength >=4
2 Strength >=3
3 Strength <3

XI ***Finger radial extensors***
0 Strength 5
1 Strength >=4
2 Strength >=3
3 Strength <3

XII ***Triceps***
0 Strength 5
1 Strength >=4
2 Strength >=3
3 Strength <3

XIII ***Deltoid***
0 Strength 5
1 Strength >=4
2 Strength >=3
3 Strength <3

B) FATIGABILITY / FUNCTION

I ***Ptosis***
0 No fatigability
1 Ptosis appearing or worsening between 30" and 60" of Simpson's test
2 Ptosis appearing or worsening before 30" of Simpson's test

II ***Upper limb girdle*** *(in the sitting position)*
0 Holds 90° abducted arms for more than 120"
1 Holds 90° abducted arms for more than 60"
2 Holds 90° abducted arms for more than 30"
3 Holds 90° abducted arms for less than 30"

III ***Lower limb girdle*** *(supine)*
0 Holds 90° flexed hips and knees for more than 120"
1 Holds 90° flexed hips and knees for more than 60"
2 Holds 90° flexed hips and knees for more than 30"
3 Holds 90° flexed hips and knees for less than 30"

IV ***Chewing*** *(Read the question (0) to the patient and assign score 0 if the answer is negative; if the answer is affirmative, ask the patient to choose which of the three statements most closely matches his condition)*
(0): Did you have any difficulty in chewing during your last meal ?
(I): My last meal lasted longer than normal because my chewing got weak.
(II): During my last meal chewing got weak after a few chews.
(III): During my last meal I was not able to chew at all.
0 Normal
1 Late fatigability
2 Early fatigability
3 Ineffective / Impossible

V ***Tongue*** *(The patient is required to press the tongue against the cheek five times on each side alternately and the examiner evaluates the force. Score 1 is assigned when a strength reduction of at least one MRC point is observed between the first and fifth time)*
0 No fatigability
1 Fatigability

VI ***Swallowing*** *(Read the question (0) to the patient and assign score 0 if the answer is negative; if the answer is affirmative, ask the patient to choose which of the three statements most closely matches his condition)*
(0): During your last meal were you able to swallow normally both solids and liquids?
(I): At the end of my last meal I had problems swallowing.
(II): I had problems swallowing early during my last meal and was not able to eat all the types of food offered.
(III): During my last meal I was not able to swallow.
0 Normal
1 Late fatigability
2 Early fatigability
3 Ineffective / Impossible

VII ***Voice*** *(the patient counts from 1 to 20 as loudly as possible)*
0 Normal; no changes.
1 Late fatigability (gets nasal or feeble after 10)
2 Early fatigability (gets nasal or feeble before 10)
3 Nasal or feeble from the beginning

VIII ***Breathing***
0 Normal
1 Slight reduction in expansibility, but normal respiratory frequency
2 Reduction in expansibility, tachypnoea and/or intervention of auxilliary muscles
3 Mechanically assisted ventilation

IX ***Neck flexors*** *(If strength according to MRC = 3 assign 2)*
0 20 flexions possible
1 fatigability within 20 flexions
2 fatigability within 10 flexions

X ***Squats*** *(hold patient by both hands without pulling)*
0 Gets up more than 10 times
1 Gets up 5 to 10 times
2 Gets up 1 to 5 times
3 Does not get up

MG disability scale

For each item, choose the score that most closely matches your condition during the last 24 hours.

1) The strength in my arms and in my hands enabled me to perform normal activities, such as brushing my hair, lifting objects, putting on my sweater, shaving, putting on make up, etc.
0 without any problem
1 with slight difficulty, or needing a longer time than usual
2 with great difficulty
3 practically prevented me from performing such activities.

2) The strength in my legs enabled me to get up from bed and chair, to go up the stairs etc.
0 without any problem
1 with slight difficulty
2 with great difficulty or with support needed
3 practically prevented me from performing such activities.

3) my ability to eat and drink (chewing, swallowing etc.) was:

- *0 completely normal*
- *1 had slight difficulties*
- *2 I had great difficulties*
- *3 I was practically unable to eat and drink*

4) My voice was

- *0 completely normal*
- *1 I had slight and transitory difficulty speaking because my voice got nasal or feeble*
- *2 I had great difficulty speaking*
- *3 I was practically unable to speak as my voice was unintelligible*

5) My sight was:

- *0 completely normal*
- *1 slightly and temporarily hampered because of drooping lid and/or double vision*
- *2 greatly hampered because of drooping lid and/or double vision*
- *3 I was practically unable to read because of drooping lid and/or double vision*

6) My breathing was:

- *0 completely normal*
- *1 I was slightly short of breath during exercise*
- *2 I was very short of breath, even at rest*
- *3 I needed breathing devices*

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